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AUTHOR Cohen, Donald J.; And Others
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ABSTRACT

The general nature of neuropsychiatric disorders of childhood, and the areas in which gains in understanding such disorders have been made are discussed in the first portion of this paper. Thereafter, discussion focuses on the contemporary approach to neuropsychiatric disorders of childhood as exemplified by three serious disturbances: attention deficit disorders, autism, and Gilles de la Tourette's syndrome of chronic multiple tics. Central processing or cognitive difficulties, attentional and activity regulation, personal/social/motivational disturbances, individual assessment, and treatment related to attention deficit disorders are described. The discussion then moves to delineate the characteristics of autism in the early and later years of childhood, as well as the causes and treatment of autism, and prospects for increased understanding of autism in the future. The discussion of Tourette's Syndrome focuses on the nature, causes, and treatment of the disorder. In a concluding overview, it is indicated that perhaps the predominant characteristic of the past several years of clinical investigation of neuropsychiatric disorders of childhood is a steady progression of knowledge and recognition of the complexity and interrelationships of developmental disorders. (Author/RH)

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MATRIX NO. 11B

NEUROPSYCHIATRIC DISORDERS OF CHILDHOOD

by

Donald J. Cohen, M.D.
Professor, Pediatrics and Psychiatry
Child Study Center
Yale University
Co-director, Yale Mental Health
Clinical Research Center

and

Barbara K. Caparulo, M.S.
Associate in Research
Child Study Center
Yale University

and

Bennett A. Shaywitz, M.D.
Associate Professor
Neurology and Pediatrics
Chief, Pediatric Neurology
Yale University

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Washington, DC 20013

NEUROPSYCHIATRIC DISORDERS OF CHILDHOOD

From the Child Study Center and the Departments of Pediatrics, Psychiatry, and Psychology, Yale University, New Haven, Connecticut

The neuropsychiatric disorders of childhood are a cluster of serious developmental disabilities, such as autism, attentional disorders, cognitive and linguistic processing disturbances, and atypical development. These disorders emerge during the first years of life, tend to persist into adulthood, and cause enormous burdens for children and families. During the past decade, there have been important advances in understanding the biological correlates, natural history, and approach to their intervention; as yet, there are no cures or preventions. Perhaps the most impressive change in relation to these disorders during the past decade has been one of attitude. Two generalizations have become increasingly accepted. First, these disorders reflect the interaction between biological and experiential factors, with different weightings of biology and experience for different syndromes and for different aspects of a particular disorder; and second, further understanding will depend on the systematic application of methods from various disciplines. In the place of doctrinal disputes — nature vs. nurture, psyche vs. soma, psychotherapy vs. education, environmental treatments vs. biological — clinicians and investigators now are focusing new methods of study on developmental disorders and are putting hypotheses to empirical, rather than ideological, tests.

The early-onset neuropsychiatric disturbances are not rare conditions. Taken as a group, perhaps 1% of all children are afflicted with serious difficulties that will interfere with their educational, social, and vocational capacities. The costs to families and to society are extremely high. For special education in a day program, tuition may range from $8,000 to $12,000 annually. For residential programs for the most severely handicapped children, costs may reach $35,000 a year, or higher. Multiplied by the number of years in a normal life span, and the *hidden* costs to parents and siblings, the social expense of a serious disorder, such as autism, is awesome. Yet, in comparison with these costs and in contrast with other important medical conditions afflicting even fewer children, such as childhood cancer, the national investment in research for the serious developmental disabilities has continued to be minute.

Gains in understanding the neuropsychiatric disorders have arisen from the following areas:

1. Diagnosis. The development of broadly accepted diagnostic criteria has allowed clinicians and investigators to communicate their observations more clearly. The American Psychiatric Association's *Diagnostic and Statistical Manual (DSM-III)* (1980) has become the standard diagnostic reference because it is based on observable, reliable behavioral criteria. In the future, these diagnostic conventions will require empirical testing to determine their validity and to delineate biological and psychological subtypes.
2. Natural History. The difficulties experienced by children shift over the course of childhood and adolescence; knowledge of the natural history of

neuropsychiatric disturbances has increased the clinician's ability to trace underlying dysfunctions and guide treatment planning.

3. Genetics. For some disorders, the application of new methods of genetic analysis has increased our understanding of one form of biological predisposition; the further elucidation of genetic contributions may provide further keys to underlying biological factors that may guide remediation.

4. Neurochemistry. In the central nervous system, neurons communicate through the release and metabolism of small molecules (neurotransmitters) that serve as chemical messengers. Many compounds now have been identified, and clinical investigators can trace the synthesis, release, and metabolism of neurotransmitters in children through assays of various bodily fluids. Basic studies of brain metabolism have provided a new picture of brain function with important, heuristic implications.

5. Cognitive Processes. Studies of language development have traced the origins of a child's first language and the formal systems that govern language elaboration; the study of language, in turn, has been related to other cognitive processes. These methods – when applied to children with neuropsychiatric disorders – have highlighted the specific ways in which learning and language-disordered children are handicapped and the relations between such disorders as childhood autism and childhood aphasia.

6. Social Development. Infants are naturally most interested in their caregivers, and humans mediate the child's experiences of the world throughout development. It is upon their motivation to understand the human world of activities that infants map their other cognitive processes, and the structures of thinking manifested in language. The scientific recognition of the close ties between affective (emotional and social) and cognitive development has helped bridge the gap between investigators who have emphasized one area at the expense of the other.

7. Intervention. Gone are the days when clinicians thought of one disorder-one treatment. Instead, multiple approaches to intervention are recognized as optimal treatment for most disturbances. In such a strategy of intervention, clinicians, educators, and parents collaborate in designing programs that may include special education, behavior modification, psychotherapy, parent guidance, group experiences, and the judicious use of medication.

In short, the past decade has seen the wisdom of the broad-gauged clinical approach in which children are seen as psychosomatic *wholes* living in the complex world of parents and others, and in which the approach to intervention must take into consideration a range of possible contributing factors and a range of possibly helpful treatments.

The contemporary approach to neuropsychiatric disorders of childhood will be exemplified by three quite different, serious disturbances: attention deficit disorders, autism, and the syndrome of chronic multiple tics of Gilles de la Tourette. Our understanding of each of these syndromes has changed dramatically during the past years; each, however, remains without known etiology and in need of many more years of systematic investigation.

Attention Deficit Disorders

In virtually every elementary school classroom, there are two or three children, usually boys, whose behavior can be described in the following terms: hyperactive, distractible, impulsive, poor reader, irritable, moody, slow learner, inattentive, dreamer, underachiever. Simple labels, such as minimal brain dysfunction (MBD), hyperkinetic child, learning disabled, hyperactivity syndrome, and tension discharge syndrome, may obscure the need for careful diagnosis and differential planning. Most recently, children with this behavioral presentation have been diagnosed as having Attention Deficit Disorder (ADD). Careful evaluation of such children will reveal a complex array of neuropsychiatric difficulties.

Developmental Sectors

Central processing or cognitive difficulties. Most children who are medically evaluated by pediatricians and child psychiatrists for learning and attentional problems do not have an even or global reduction in intellectual ability. Their cognitive difficulties affect one or more specific areas of central processing, such as: (1) coding and decoding symbols (particularly words); (2) scanning complex displays; (3) detecting auditory or visual signals (messages) embedded in distracting sounds or displays (noise); (4) rapidly arranging symbols with accuracy; (5) generating a meaningful whole from auditory or visual cues; (6) analyzing passages in writing or speech into smaller units; or (7) assembling individual units into sequentially ordered paragraphs. Difficulties in cognitive and perceptual analysis are unmasked most clearly during the first grades of school, as the child is faced with academic tasks that demand organization of symbolic representations.

Attentional and activity regulation. Attention is a multifaceted concept, covering several associated processes: maintenance of a conceptual set or disposition for action; vigilant readiness to respond to the occurrence of a stimulus condition; perseverance on a task; orientation to an event or array; or a state of general expectancy. The ability to sustain an external focus of attention (e.g., on a teacher's statements) is connected closely with the capacity for maintaining an internal focus of attention on thoughts (e.g., to remember the plan of action or intention). Classroom and home activities call upon all aspects of attentional regulation: listening and looking, blocking out extraneous stimuli, remembering instructions, isolating aspects of a story or problem, and being ready to end one line of action or thought and begin another.

Children with attentional problems move from one site to another, constantly diverted by sounds and objects; they are unable to inhibit action or to await a stimulus that signals activity. Thus, their behavior is chaotic and not in context. Both at home and in school, children with attentional difficulties evoke a great deal of anger and often are bewildered when they are scolded.

The regulation of attention is associated closely with the modulation of activity. In contrast with children endowed with normal attentional mechanisms, those with disturbances in concentration do not have their attention captured by the normal range of stimuli, and they stay fixed for shorter periods of time. Motor activity, thus, is less inhibited: when other children are silently watching a movie, impaired

children are flooded with action and ideas. While attentional problems are perceived most vividly when they are accompanied by gross hyperactivity, a few children with attentional difficulties are generally hypoactive daydreamers. The troublesome, active child often precipitates a vigorous response from his teachers early in the school term; the hypoactive, overly good, compliant child with an attentional problem may drift from grade to grade before his teachers and family recognize that s/he has a serious problem interfering with school achievement.

Motor control. Major disturbances in movement – such as cerebral palsy – and major problems in fine motor control are readily apparent to parents and physicians. Most children with school learning and behavior problems do not have any findings of this magnitude, nor do they have any other evidence of structural brain dysfunction manifested on the neurological examination, skull X-ray, or electroencephalogram (EEG). Instead, they more frequently present with a range of motor atypicalities or immaturities that are generally labeled "soft" or "minor" neurological signs. This is a bad designation, because these signs are generally neither unimportant nor only subjective.

Neuromaturational difficulties in motor control may be apparent from careful observation of the child at play and while working on various kinds of tasks. Formal testing may be needed to elicit or provoke certain impairments for which the child, especially if s/he is intelligent, may have learned to compensate. That formal neurological examination may be totally normal or reveal only a medley of findings suggests a diffuse dysfunction in central nervous system maturation (particularly in relation to areas concerned with motor regulation and inhibition) and a resultant disturbance in the formation of coherent, stable body schema. Children with *motor control disturbances* generally are aware of their handicap and of the reason for exclusion from sports and their poor grades in penmanship.

Personal-social-motivational disturbances. Children with difficulties in school achievement, attention, and motor control may have any type of personality difficulty, or none. Those who are aggressive, with peers or adults, who constantly *test* rules and who appear impermeable to criticism and punishment frequently are referred for psychiatric evaluation and are perhaps most likely to receive punitive treatment by schools or active medical intervention by physicians. However, other children who suffer from childhood depression – lack of pleasure or initiative, social inhibition and withdrawal, low spirits, fearfulness, and hypochondriacal concerns – may have their personal difficulties overlooked. Children may place themselves into situations of actual danger or attract attention by constant clowning, perhaps in reaction to underlying fears; as school problems exacerbate, they move into more organized groups in which such life patterns are socialized. At times, families may describe a ruthless, thoughtless, overly active child who turns out to be a nice youngster victimized by compulsive, depressed, overly perfectionistic, or maritally upset parents who need the child as the ticket of admission into treatment, as the camouflage for their own dilemma, or as the escape valve for their tensions.

Individual Assessment

The diagnostic assessment provides information about the child's cognitive processing, attention, academic achievement, personal relations, reality orientation, anxiety level and methods for its modulation, neuromaturational level, family life, school experiences, and community. The initial chief complaints – hyperactivity, school failure, belligerence – become embedded in a formulation that describes the child's genetic, physiological, and experiential history in the context of family, school, and community. This formulation rarely can assert the cause of a child's difficulties, because the lives of children are not subdivided into those neat categories that guide the writing of textbooks or the organization of university departments. Often, the clinician considers various contributory factors. Prematurity, birth anoxia, difficult delivery, or evidence of gestational problems (e.g., staining) may orient the clinician to neurological or maturational insult early in development for a small group of children. This area of insult, however, once so highly emphasized in the hypothesis of continuum of reproductive casualty (which held that mild trauma led to mild deviations and more serious birth problems, to epilepsy and mental retardation) now no longer is seen as playing a major role for the great majority of children with developmental difficulties. Similarly, there is rarely a single medical lesion – such as meningitis, head trauma, or encephalitis – or sensory problem to which subsequent learning problems may be traced. Instead, the clinician will balance the child's endowment, vagaries of early experiences and moves, parental style, school environment, community stresses, and the like, and arrive at a weighting for each sector. Some children will be seen as having inborn disturbances in attentional and anxiety-regulating mechanisms, perhaps associated with neurochemical dysfunctions; others, as carrying the scars of neglect and abuse, perhaps at the hands of parents who superficially seem like quite solid citizens; and for others, the child's innate endowment and disposition and the parents' capacities will be seen to have been badly matched. Many others will be found to be responding to transient or prolonged environmental burdens, at home or in school.

Treatment

Careful diagnostic assessment will suggest: (a) what types of interventions are appropriate, (b) which are available, and (c) what therapeutic strategy is most likely to be useful.

Parental guidance is aimed at helping the parents to understand the child's problems, developing better approaches to helping the child, and explicating what other forces may be contributing to the child's problems.

Special education. The explication of special areas of learning difficulty or the interference with learning because of prolonged motivational problems can lead to the design of special educational interventions. Most schools, and parents, wait far too long before moving a child from the regular classroom into special *learning laboratories* for part or all of the day. Remedial tutoring, special precision education methods, and small, structured classrooms should be considered as soon as the child's academic and behavioral difficulties in the classroom are recognized, not in the third or fourth grade when he is seen by his classmates and by himself as *bad* or *dumb*.

Psychotherapy. Anxious, inhibited children and children whose aggressive behavior reflects intrapsychic conflict may respond to conventional child therapy, and school and other behavioral problems may be resolved over the course of months.

Physical education. Poorly coordinated children with gross disturbances in body schema usually become observers in gym classes, although they need such programs far more than athletic peers. Gym teachers often may be helped to recognize these children as special challenges, and *modern* physical educators can evaluate expertly motor skills and design special programs.

Medication. Over 600,000 children are receiving stimulant medication in the United States, and we are one of the few countries where this medication is used at all. In England, consultant psychiatrists may prescribe methylphenidate to a few children annually. In the United States, there are physicians who maintain hundreds of children on such drugs. Various theories have been proposed, including the use of medication to suppress children, to avoid real school reform, or to profit the prescribers and the manufacturers. The enthusiastic use of medication also may reflect quite different forces: better case finding; higher incidence of learning and behavioral disturbances (associated with the pace of life, in general); and increased expectations and the unwillingness in our society to label children as slow learners and to restrict them to nonacademic courses, for examples. The truth probably lies somewhere between those who advocate a trial of medication for all behaviorally disturbed children and those who would deny such treatment to any.

For the child with specific difficulties in attention and concentration, stimulant medication may produce almost immediate improvement in school learning, social performance, and pleasure in family life. With improved attentiveness, diffuse hyperactivity decreases and the child seems better able to appreciate the normal controls of his environment. After a period of time, the medication may be reduced and finally withdrawn, and skills in school and life may persist. For most children, the use of stimulant medication must be accompanied by other types of intervention, including behavior modification, special education, and family counseling.

Early Childhood Autism

In 1943, Leo Kanner identified a small group of children with a profound inability to establish meaningful emotional and social relationships, even with their parents, and who failed to develop language in the normal way. He characterized this group as suffering from *early infantile autism*, a disorder that he considered to be an inborn vulnerability, similar to inborn errors of metabolism, such as phenylketonuria (PKU). The choice of the term "autism," suggested by the autistic withdrawal found in adult schizophrenia, was perhaps unfortunate. Over the past 40 years, research and clinical experience have demonstrated that autistic children have multiple cognitive and emotional handicaps that block acquisition of basic concepts of self, other, and mutuality, and prevent recognition of the symbolic nature of language. When, later in life, autistic youngsters appear to withdraw from social contact, it is an expression of confusion and anxiety related to overly complex social demands rather than a retreat into the internal world of fantasy. Contemporary studies of autism continue to search for the biological basis of the disorder; analysis of the dys-

functions of autistic children has revealed disturbances in the preconditions for normal cognitive and emotional development, and disruptions in the processes and skills emerging in the first year or two of life upon which later linguistic and social competencies depend.

Autism occurs in about 1 in every 2,500 children, making it far more common than PKU (1:15,000), and less common than Down's syndrome (1:700). It affects three or four times more boys than girls, and, while it is unusual for more than one child in a family to have autism, there are families with autism, language disorders, and other developmental disabilities in siblings and relatives that suggest some genetic contribution. Because of its severity and the continuing uncertainties surrounding its causes, diagnosis, and treatment, childhood autism has generated a great deal of scientific research, discussion, and treatment approaches.

The Early Years

The behavioral and emotional disturbances of childhood autism usually are apparent from the first months of life. Failure to establish or maintain eye contact, to prepare for being picked up, and to smile socially are among the first symptoms of the disorder. The baby may be unusually good or cry inconsolably; he may become preoccupied with one object or toy, spend hours looking at his fingers or repeatedly banging his head against his crib. The child's physical health during the first year of life is, typically, normal, although feeding difficulties, marked by strong preferences for only some foods, are common. Abnormal sleeping patterns, allowing the child and his family only 3 or 4 hours of sleep a night, often are reported. Activity level appears to be poorly modulated; extreme hyperactivity and overarousal to environmental stimulation mark the early years for many autistic children, although others show a hypoactive behavioral pattern and appear sluggish and almost apathetic to what goes on around them. The child often seems not to hear the speech addressed to him, but panics at the sound of a washing machine or hair dryer. The most perplexing, and, for parents, devastating aspect of the young child's development, however, is the discrepancy between his gross motor and physical development and his growth in social and linguistic skills. The child's inability to relate to other human beings in the normal way becomes more noticeable as he grows older. Parents may be used as objects in order to satisfy basic needs, but the child shows no signs of closeness and enjoyment from mutual interaction with his parents. Physicians, finding the child's physical health and motor development to be normal, have typically dismissed parents' expressions of concern or have suggested a *wait and see* policy. By the age of 16 to 24 months, however, the child's history of failure to babble, play social games like peek-a-boo, and learn single words results in referral for developmental evaluation. Careful assessment of the preschool autistic child reveals a range of intellectual, linguistic, social, and behavioral abnormalities, each suggesting a single diagnosis such as mental retardation, developmental language disorder, atypical personality development. and severe emotional disturbance. While each label captures a particular facet of autism, none fully describes the range of deficits present in the disorder.

The Later Years

Research is only just beginning to fill in the gaps in the natural history of childhood autism by studying the children, once physically attractive and motorically agile, as they move through middle childhood, adolescence, and into their adult years. Only 5% to 15% of older autistic persons develop useful communicative language and improved social and work skills that allow them to live in semiindependent or, rarely, independent, situations. Their behavior in social encounters, while not bizarre or disruptive, lacks spontaneity and reflects the hard work they, and their parents and teachers, have put into their education. In school, these persons show areas of high intellectual ability whose utility in terms of acquisition of practical skills, however, may be questionable. After extreme delays in learning basic linguistic structures, the higher functioning autistic person becomes quite competent in using language, but experiences major difficulties in understanding the social significance of interpersonal communication. As an adult, he appears anxious, fearful, and often depressed about his disabilities; when upset or excited, especially by social demands, he often engages in the stereotypic behaviors that first appeared years earlier. Friendships and emotional ties with adults remain superficial, despite his feelings of recognition that such relationships would help to ease his loneliness and sense of isolation.

For the less fortunate autistic child whose language does not progress, behavior during the school-age and adolescent years remains similar to that of the preschool years. Hyperactivity may decrease with maturation and training, but the child's ability to communicate, use symbols, follow commands, and relate to peers and adults is extremely limited. Some children become more aggressive toward others and destructive, symptoms that require thoughtful but firm attention through behavior modification. As the child becomes older, his intellectual and linguistic handicaps may become more obvious so that by the age of 17 or 18, they overshadow his social impairments. For the autistic child with greater intellectual impairments, intense efforts must be made to prevent stereotypic behaviors from dominating his waking hours, by redirecting his attention toward basic skills and comprehension of simple instructions.

The upsurge of sexual drives during adolescence may lead to increased tension that the autistic man or woman does not know how to discharge. For some autistic individuals, parents and educators must consider teaching even basic sexual knowledge, such as how, when, and where to masturbate. The sexual curiosity and inappropriate behavior toward others lead to difficulties in school and residential programs. The autistic person cannot fully understand the feelings of others or navigate the subtle mutualities required for normal sexual relations; his normal sexual urges thus can find no socially acceptable avenue for expression. Curricula for sex education of autistic children recognize these special problems and suggest methods for teachers, parents, and the individual to deal with them.

Causes of Autism

The cause of primary childhood autism is not known. Psychoanalytic and psychodynamic theories, which were most prominent in the 1940s and 1950s, have been rejected in favor of hypotheses emphasizing an organic basis of the disorder. In

support of such biological theories are various kinds of evidence. Disturbances in sensory integration and in the sequential ordering of behavior appear to reflect abnormalities in central nervous system functioning. Autistic children's stereotypic behavior, disorganization, hyperactivity, troubles in feeling pleasure or pain, and other symptoms are consistent with some patterns of abnormalities that affect brain metabolism. Many autistic children have an abnormal or borderline abnormal brain wave pattern, and about 25% develop seizures. Autism is found throughout the world, in every social class, with a uniform clinical picture. And finally, in a high percentage of cases in which one identical twin has autism, the other also has this condition, suggesting a congenital, and perhaps genetic, basis for the disorder. All of this evidence supports the view that autism is like some of the now well-defined inborn errors of metabolism, but considerably more research is needed to investigate such an hypothesis.

Another promising field of investigation is the possible biochemical basis for severe developmental problems. Recent research on manic-depressive disease and schizophrenia in adults, as well as on neurological diseases such as Parkinson's syndrome and chronic multiple tics of Gilles de la Tourette's syndrome, has greatly increased knowledge about the way in which messages are chemically transmitted by nerve cells in the brain. This process involves various chemicals – such as dopamine and norepinephrine – which are stored in nerve endings and then released when needed. There may be abnormalities in the way in which these chemicals are synthesized, released, or broken down after performing their function. Knowledge about these processes has been the bases for the treatment of Parkinson's disease with DOPA and for explaining the chemical action of medicines used in the treatment of depression, schizophrenia, and childhood attentional disorders.

Treatment

Just as the cause of autism is not known, its cure remains undiscovered. The mainstays of treatment are special education and vocational training, parental guidance, psychotherapy, drug therapy, and a structured, consistent living environment.

Educationally, the major advances in recent years have been in the use of behavior modification integrated with cognitive-developmental curricula, beginning in the preschool years and extending through to adulthood (see Rutter & Schopler, 1978, for review). Parents are taught to educate their own children, even at the age of 2 or 3 years, during the hours spent at home. Behavior modification is an effective method of both training the child's behavior and teaching him self-help, preacademic, and academic skills when used in the context of precision education, in which the child's capacities, the teacher's-parent's objectives, and the teaching and measurement methods are precisely defined. Both educational techniques, however, are extremely complex and have been abused by even well-intentioned educators; their application requires sensitivity, experience, intuition, and intelligence, and their success depends on the establishment of close collaboration between parents and professionals who can maintain constancy from home to school. Their utility is increased when used in conjunction with a comprehensive language and cognitive development program aimed at providing the child with a range of concepts about objects, people, the world, and causality.

Prevocational and vocational training must be begun as the child approaches early adolescence. The development of a work, self-help, and social vocabulary, the acquisition of a range of vocational skills, and the learning of on-the-job behaviors (e.g., paying attention, keeping track of one's time spent at work, following instructions) are the three primary areas of training that must be addressed in a step-by-step, consistent way. For better functioning persons with autism, the acquisition of vocational skills provides a channel for partial self-support as well as for assuming an acceptable role in society, and makes a critical difference in ultimate prognosis. For autistic persons who are more impaired, prevocational and vocational training offer the opportunity of participating in highly structured and well-supervised workshops where social and intellectual growth may continue well after formal education ends. The prognosis for severely impaired persons with autism remains guarded, although models now exist of adult living environments that offer reasonable alternatives to a life spent in an institution.

Parents require emotional support and practical, long-term guidance in relation to both their autistic child and their other children. An autistic child may create extreme tension between a husband and wife, which, when coupled with exhaustion, anxiety, and guilt, ultimately may lead to separation and divorce. Counseling siblings of an autistic child is a critical part of the supporting professional's job, as is aiding in the development of a life plan for the child that allows the family to respond rationally to critical changes accompanying developmental transitions (e.g., preadolescence, entry into adulthood).

Psychotherapy plays a limited role for most children with autism, but for those who show the beginnings of being able to form social and affectionate attachments, and for some older, better functioning youngsters, it offers the opportunity of expressing feelings of anxiety, happiness, and self-discovery. There are no firm data to support this belief, but several youngsters, studied through late adolescence and into adulthood, have appeared to benefit from the personal contact and feelings of relatedness they have experienced in therapy with an empathetic and intuitive professional.

Although no psychoactive medication has proven to be especially useful for all symptoms or for long periods of time in autistic children, judicious clinicians may suggest a trial of a medication with demonstrated value in the treatment of severe psychiatric disturbance, hoping to relieve a target symptom, such as extreme self-aggression, or to help a child make use of special education by relieving his hyperactivity or anxiety. Children should not be kept on medication for years, however, and close monitoring of the dose and side effects by a responsible physician, the child's parents, and his teachers is essential. Medication such as dextroamphetamine, which has traditionally been used in the treatment of hyperactivity, has not been found to be useful with autistic youngsters, since it increases the behavioral and mental disorganization the child experiences and appears to exacerbate other symptoms.

Finally, children with autism require continuity of care and stability in their lives. Residential special education programs often provide the best setting to meet these needs because of their ability to have 24-hour staffing by persons who can respond

consistently to the child's behavior and emotional needs. The child's place within his family, however, must not be sacrificed; weekly or monthly visits home help the family remain engaged with the child. Alternatives to residential education include *extended day* programs with the local school system or in private schools, such as Benhaven, where vocational, academic, and social training is offered on an 8-hour day, 6-day week, 50-week year, basis. Summer *vacations* should be avoided, since serious regression almost always occurs, and the child's educational needs go unattended. Finally, autistic children should not be placed on general wards in institutions for the mentally retarded or seriously emotionally disturbed; they invariably do badly, lose areas of competence, and become more disorganized behaviorally.

Future Prospects

Until the causes of autism are understood many children and their families face a long and exhausting course in trying to attain the diagnostic, educational and treatment services that have been developed over the past 20 years. Perhaps the most exciting advances of the 1970s have been in the elucidation of the kind of educational programs that most benefit persons with autism. In the future, interdisciplinary and parent-professional collaboration must take the lead in assuring that such programs reach those who need them. Great work remains to be done in establishing adult living environments for persons with autism, and in assisting families in planning for the long-term needs of their autistic child. Groups, such as the National Society for Autistic Children, now play a central role in advocacy on behalf of persons with autism and similar developmental disorders.

Research must continue in brain chemistry and physiology, both in relation to primary autism and those developmental disorders, such as central language disturbances, that may be related, and to the changes in brain functioning over the natural course of these disorders.

The development of social policies about the treatment of autistic persons, the improved provision of services, and the continuation of biological research requires an intense interdisciplinary effort and collaboration with parents.

With the expansion of knowledge in the neurosciences, linguistics, education, and other areas relevant to autism, sustained collaboration between professionals and parents promises to reveal new insights into the causes of, and treatments for, severe childhood disabilities.

Tourette's Syndrome

Tourette's syndrome, named after the 19th century neurologist who first described it formally, was once thought to be a rare, exotic disorder; it is now a well-defined neurophysiological condition which is probably more common than childhood autism. Tourette's syndrome is important as a serious developmental disability in its own right; it takes on added relevance as an exemplary syndrome for understanding the interaction between biological predisposition, clinical characteristics, and various pharmacological agents.

Tourette's syndrome (TS) occurs predominantly in boys (with a ratio of 4 to 8 males for every girl) and presents during the first years of life as attentional and behavioral difficulties quite similar to those described earlier in our review of attention deficit disorders. Between ages 4 and 16 years (with a mean age of onset of 7 years), the child develops his first motor symptoms. These usually consist of simple facial tics (e.g., eye blinking or head jerking), and the symptoms progress to increasingly complex, diverse movements affecting virtually any muscular group in the body (arm thrusting, tongue protrusion, hopping, spitting, and the like). Phonic tics appear soon after motor ones and may include simple noises followed by interruptions in the flow of speech, hesitancy, repetition, and inappropriate outbursts of words, which are sometimes obscene. The child may feel that he is losing control as he sees himself doing and saying things that are inappropriate and totally out of character; parents panic as they see their bright, beloved youngster become increasingly distracted and driven by his incessant need to emit movements and sounds. Symptoms change over time, and periods of relative remission may be followed by longer periods of exacerbation. In addition to movement difficulties, perhaps a majority of patients with TS have learning disabilities, school problems, and compulsive behavior. In addition, behavioral disorders and depressive conditions may be associated with the negative social interaction elicited by the symptoms or may reflect aspects of the underlying pathobiology.

The symptoms of TS stand between the mind and the body. The underlying disturbance in TS appears to be a neurochemical or physiological dysfunction in the way in which impulses are inhibited in the central nervous system. Action tendencies, thoughts, movements, and impulses that normally are inhibited are, in TS, expressed in conscious thought and action. Sometimes, the movements seem to be emitted with no prior warning; more often, and perhaps in most cases, the child is aware of a growing tension in a part of his body or a strengthening impulse to do or say something. He may try to resist the action or sound, just as a person with a habit may try to hold it back; however, after a shorter or longer time, he feels the irresistible urge to perform the tic and can no longer hold it in. The release of the movement or sound is followed by a reduction in tension, only again to be followed by the increasing pressure for discharge. Thus, children and adults with TS feel that they are controlled by their own impulses.

TS may be a lifelong disorder and, when it is severe, may pose extreme limitations on an individual's educational and work choices. Yet, despite the obvious burden on social relations posed by Tourette's, many individuals with the syndrome manage to earn higher educational degrees and to become productive members of society.

Causes of Tourette's Syndrome

As with the other neuropsychiatric disorders, the cause of TS remains unknown. However, the last several years have brought exciting gains in knowledge. Today, we know that there is a definite genetic basis for the disorder, at least in many cases. TS runs in families, and children with TS have a higher frequency of relatives with the full-blown condition or with chronic multiple tics. The expression of the underlying predisposition to TS is sex-modified: girls with TS tend to have a heavier genetic

loading. While we can trace the expression of TS in families, we do not know what is transmitted genetically.

Various lines of evidence have suggested that central nervous system metabolism is disturbed in TS. During the 1960s, Dr. Arthur Shapiro demonstrated that TS responds very well to haloperidol, a potent neuroleptic medication. During the past years, this observation has been repeated with hundreds of patients who have experienced dramatic improvement with haloperidol. Since haloperidol reduces the functioning of the dopamine system in the brain, it was hypothesized that TS was an expression of an overactive dopamine system. Studies of cerebrospinal fluid and other research in patients with TS have supported the idea that dopamine regulation may be involved in TS. During the past several years, however, investigators have found possible disturbances in other neurochemical systems, such as the norepinephrine, serotonin, and acetylcholine system in patients with TS. The possibility of studying neurotransmitters in patients is one of the amazing advances in clinical research in childhood neuropsychiatry. Along with the neurochemical observations have come suggestions for new approaches to treatment. While haloperidol sometimes is extremely effective, it is not useful for all patients. Some patients do not respond well to it from the outset; others develop serious side effects (such as movement difficulties, phobias, depression, and intellectual dulling). Other medications, such as clonidine and pimozide, have been used for some of these patients, with good results.

Treatment

It is now clear that children with TS often require more than a medication. Because at least 50% of the children have school learning difficulties, they generally require special tutoring or programs of special education. Behavioral problems that arise in association with TS, or because of social reactions to the syndrome, need to be addressed by therapy and special groups; and the parents and siblings of children with TS need guidance and support. As the genetics are further elucidated, there may be a role for genetic counseling and attention to the special concerns that this will generate in families and children.

Thus, at the present time, investigators are engaged in multifaceted approaches to TS. They are trying actively to explicate the genetic contributions, the underlying neurochemical disturbances, and the related attentional, cognitive, and learning difficulties. Clinicians are aware of the need for systematic, multimodal treatment programs. While the most intensive commitment of clinical investigators is to find specific cures, there is the clear recognition that a good deal already can be done for individual patients. Finally, the recognition that medications always carry with them serious side effects has led to increased caution. For example, the possibility that haloperidol may lead to tardive dyskinesia (a sometimes irreversible movement disorder), intellectual dullness, phobias, and other biological and psychological side effects has increased the motivation of clinical investigators to find other, alternative forms of treatment. Children with TS are seen not simply as having unusual movements, but as whole people in need of broad-gauged therapy.

Just as with autism and learning disabilities, the strongest force in informing the public and stimulating research and new services has been a lay organization, the National Tourette Syndrome Association. From a small, parent group, the Tourette Syndrome Association has grown into a national organization with thousands of members. This organization, and its professional consultants, have brought TS into the public awareness through extremely effective use of television, newspapers, radio, and other media. TS, once considered extremely uncommon and strange, increasingly is recognized as a medical disorder afflicting children in every school district, and many adults. Families of TS patients, and the children and adults themselves with TS, have been strengthened by the knowledge that others have coped with the disorder and that there is no shame in having a medical condition. The national society has educated pediatricians, teachers, and school psychologists, who are in a position to refer children for specialized evaluation; because of education, the years of uncertainty and anguish between the onset of symptoms and diagnosis have been shortened. Thus, the Tourette Syndrome Association, the National Society for Autistic Children, the National Association for Children with Learning Disabilities, and similar parent-led organizations, have demonstrated one of the most exciting new movements in medicine – the active collaboration between consumers and professionals in the generation of new research, the provision of services, and the education of the public.

Overview

During the latter part of the 19th century and the first decades of this century, clinicians recognized a range of early onset, often persistent, serious disorders of development. Numerous categories were devised, such as autism, childhood schizophrenia, minimal brain dysfunction, hyperactive child syndrome, multiple tic syndrome, and the like. Around each of these syndromes were controversies about etiology and treatment. Out of ignorance, clinicians often accused parents of causing these disorders, when, in fact, parents were their child's most important advocates and his or her only hope for survival in a society that offered very little in the way of effective treatment. At times, other *causes* were proposed and various *treatments* were utilized – sometimes at great human and financial cost. Yet, over the past decades there has been a steady progression of knowledge and a recognition of the complexity and inter-relationships of developmental disorders. Diagnostic criteria have been clarified and natural history described. Clinicians and investigators increasingly have recognized the need for basic, systematic research, and not simply the hectic search for *the cure*. This orientation, perhaps more than any specific findings, as such, is most characteristic of the past several years of clinical investigation. The application of new methods – from biological, psychological, and other branches of inquiry – to the neuropsychiatric disorders is evidence that these disorders are being taken seriously. They are worthy of the most intelligent applications of front-line methods of inquiry, just as the other devastating medical disorders of childhood. Until cures and preventions are discovered, clinicians and investigators will have to continue the search for more effective modes of biological and educational intervention.

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